

A case of Good syndrome with refractory gastrointestinal ulcers

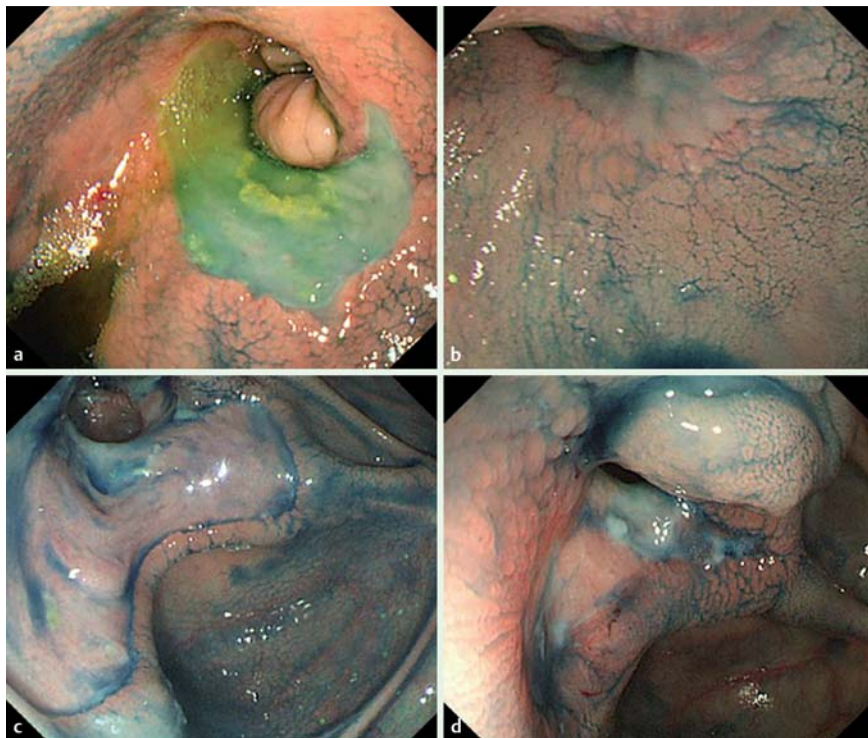


Fig. 1 **a** Endoscopic image of the stomach before intravenous immune globulin (IVIg) treatment. A shallow open ulcer is observed at the greater curvature of the stomach. **b** Endoscopic image of the stomach after IVIg treatment. A remarkable improvement of the ulcer is seen. **c** Endoscopic image of the ileocecum before IVIg treatment. A shallow open ulcer is observed at the ileocecum. **d** Endoscopic image of the ileocecum after IVIg treatment. Remarkable improvement of the ulcer is seen.

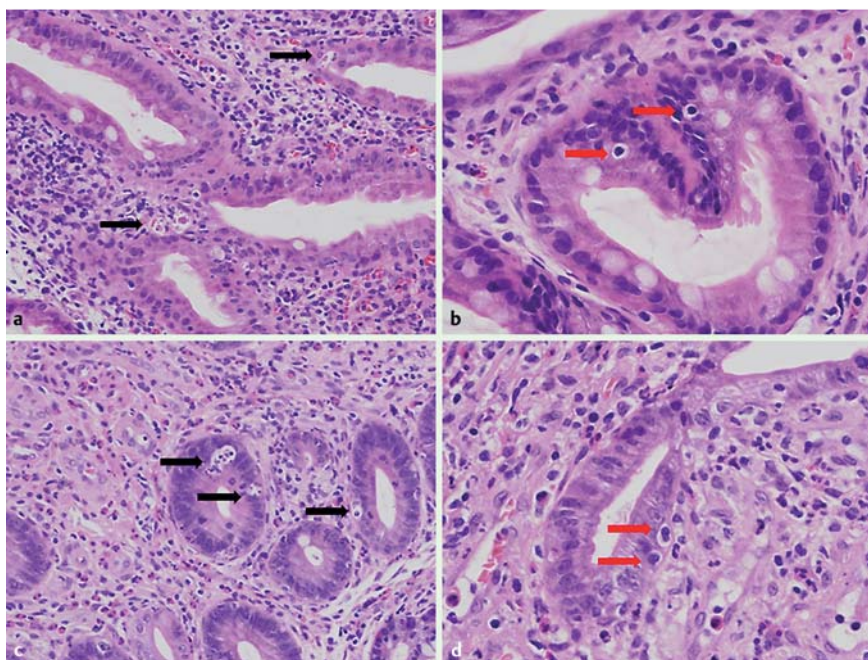


Fig. 2 **a, b** Histopathological images of the biopsy specimens of the gastric ulcer before intravenous immune globulin (IVIg) treatment. **a** Epithelial apoptosis (arrows). **b** Intraepithelial lymphocytes (IELs) (arrows). **c, d** Histopathological images of the biopsy specimens of the ileocecal ulcer before IVIg treatment. **c** Epithelial apoptosis (arrows). **d** IELs (arrows).

A 61-year-old woman was referred to our hospital in November 2011 for treatment of chronic diarrhea and refractory gastrointestinal ulcers. Her medical history included thymoma (resected at age 44) accompanied by hypogammaglobulinemia and pure red cell aplasia (PRCA), and a distal gastrectomy for gastric ulcer (refractory to proton pump inhibitor treatment and *Helicobacter pylori* eradication).

On the patient's admission, laboratory examination revealed severe hypogammaglobulinemia, absence of peripheral B cells, CD4+ T-cell lymphopenia, and inverted CD4/CD8+ T-cell ratio. Pathogenic bacteria, cytomegalovirus, and human immunodeficiency virus tests were negative. From these results and her medical history, the patient was diagnosed with Good syndrome with PRCA, and was treated with intravenous immune globulin (IVIg) every 2 weeks. At 2 months later, her immune globulin level had increased to normal. Previously upper and lower gastrointestinal endoscopy had shown an ulcer at the greater curvature (● Fig. 1 a), and similar multiple ulcers at the ileocecum and the terminal ileum on admission (● Fig. 1 c). Histopathological analysis of the biopsy specimens from these lesions showed a lack of plasma cells in the lamina propria, epithelial apoptosis, and increased intraepithelial lymphocytes (IELs) (● Fig. 2). Similarly, 2 months after initiation of IVIg treatment, these lesions were markedly improved (● Fig. 1 b, d), and the epithelial apoptosis and IELs had disappeared.

Good syndrome is defined as thymoma with adult-onset immunodeficiency, and is characterized by hypogammaglobulinemia, low or absent B-cells, CD4+ T-cell lymphopenia, and inverted CD4/CD8+ T-cell ratio [1]. Although diarrhea is the main gastrointestinal manifestation of the syndrome, in most cases no definite pathogens are identified [2]. A lack of plasma cells in the lamina propria, epithelial apoptosis, and increased IELs are characteristic histopathological findings in common variable immunodeficiency, a primary immunodeficiency similar to Good syndrome [3,4], but not in Good syndrome. This is the first report in English describing endoscopic and pathological improvement of refractory gastrointestinal ulcers in Good syndrome by IVIg treatment.

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